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Echocardiographic and Doppler Evaluation of the Aortic Arch and Brachiocephalic Vessels in Cerebral and Systemic Arteriovenous Fistulas

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Congenital arteriovenous fistulas presenting in the newborn period pose difficult diagnostic problems and simulate structural heart disease. Angiocardiography, when performed, demonstrates enlarged brachiocephalic vessels and rapid cerebral venous return. The value of echocardiographic imaging and measurement of the aortic arch and brachiocephalic vessels, and evaluation of the Doppler flow profile in these vessels as a means of making a rapid diagnosis of cerebral or thoracic arteriovenous fistula, was therefore assessed in 10 infants with these diagnoses seen over a 4 year period (1983 to 1987). Twenty-nine infants (median age 6 weeks) undergoing two-dimensional echocardiography but with no significant lesions were prospectively selected as controls.

Nine of the 10 patients had congestive heart failure at presentation (mean age 2 days). A cranial bruit was heard in three and arteriovenous fistula was suspected in five patients. Aortic arch segments and brachiocephalic vessel dimensions expressed as ratios of the abdominal aorta showed significantly larger values in patients for the ascending aorta ($p = 0.01$), innominate artery ($p < 0.001$), right and left subclavian arteries ($p < 0.001$) and left common carotid artery ($p < 0.05$). The thoracic descending aorta was, however, significantly smaller in patients ($p < 0.002$). Retrograde diastolic Doppler flow in the descending aorta proximal to the ductus arteriosus and anterograde diastolic flow with a mean spectral flow-time integral 27% of systolic were present in patients only, whereas Doppler diastolic flow in brachiocephalic vessels, present in 5 of 29 control infants, was <15% of systolic flow and not accompanied by dilation of these vessels.

The combination of one or more dilated brachiocephalic vessels, diastolic retrograde flow in the isthmus and diastolic anterograde flow in the dilated vessels is useful in reaching a diagnosis of cerebral or thoracic arteriovenous fistula, and should aid in expediting rapid definitive cerebral ultrasound and arteriography for confirmation.

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Congenital arteriovenous fistulas, most frequently involving the brain or liver, may present in the newborn period in association with intractable congestive heart failure. They often pose difficult diagnostic problems because the clinical signs may simulate both complex congenital heart disease, such as coarctation of the aorta or Ebstein's anomaly of the tricuspid valve, and nonstructural heart disease, including transient myocardial ischemia or cardiomyopathy.

Diagnosis relies mainly on the clinical finding of a cranial bruit, when audible, followed by cranial ultrasound examination for confirmation. Delay in diagnosis is therefore not uncommon because attention is focused on the heart. Angiocardiography, which demonstrates increased caliber of the brachiocephalic vessels and rapid cerebral venous return, is sometimes carried out as part of the diagnostic evaluation.

Most infants present to the cardiology service, in which the initial investigation includes echocardiography. We therefore investigated the value of echocardiographic measurements of the caliber of the aortic arch and brachiocephalic vessels, and Doppler evaluation of the systolic and diastolic flow profile in these vessels, in making a rapid and specific diagnosis of a cerebral or thoracic systemic arteriovenous fistula in a group of infants with this diagnosis.

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Table 1. Clinical Features of 10 Patients

Patient No.	Age at Presentation (days)	Cranial Bruit (+ or −)	ECG ST/T Wave Changes (+ or −)	Diagnosis	
				Initial	Final
1	6	+	−	Coarctation	av Fistula
2	2	+	−	av Fistula	av Fistula
3	1	−*	−	A-PW	RS av Fistula
4	1	+	−	av Fistula	av Fistula
5	1	−	+	Ebstein's	av Fistula
6	1	+	+	TAPVD	av Fistula
7	7	+	+	av Fistula	av Fistula
8	2	−	+	Coarctation	av Fistula
9	2	−*	−	av Fistula	RS av Fistula
10	1	−	+	av Fistula	Megalencephaly

*Patients in whom a precordial continuous murmur was heard. A-PW = aortopulmonary window; av fistula = cerebral arteriovenous fistula; Coarctation = coarctation of aorta; Ebstein's = Ebstein's anomaly of the tricuspid valve; ECG = electrocardiogram; RSAVF = right subclavian arteriovenous fistula; TAPVD = total anomalous pulmonary venous drainage. The cranial bruit in Patients 6 and 7 was heard after ultrasound imaging of the aneurysmal vein of Galen. Three patients died in the newborn period. With one exception (Patient 3), all patients were in heart failure at presentation.

Methods

Study patients. We retrospectively reviewed all the records of infants seen at the Hospital for Sick Children who were diagnosed as having cerebral or systemic arteriovenous fistula between March 1983 and March 1987. This time period coincided with the introduction of high resolution imaging and Doppler ultrasound at this institution. Of 15 patients identified, 10 had adequate echocardiographic studies. These 10 constitute the study group.

Clinical features (Table 1). The following clinical information was obtained from patients' records: age, weight at presentation and presence or absence of cyanosis, significant precordial murmurs, cranial or other bruit, cardiomegaly on chest roentgenogram and electrocardiographic (ECG) evidence of myocardial ischemia. The initial diagnosis based on clinical information alone was noted. Nine patients had congestive heart failure at presentation, seven with an arteriovenous fistula involving the vein of Galen, one with multiple right subclavian and axillary arteriovenous fistulas and one with massive unilateral hemispherical cerebral hypertrophy consisting of primitive tissue associated with marked enlargement of the left cerebral veins. At autopsy these appearances in the latter patient were believed to be consistent with increased blood flow to the left cerebral hemisphere and a primitive capillary bed.

Control group. Twenty-nine infants aged 1 to 40 weeks (median 6) with no significant intracardiac or aortic arch anomalies, who were undergoing echocardiography and Doppler evaluation for suspected cardiac disease, were prospectively selected to constitute the control group. This age range was selected to include as many neonates as possible because most infants with significant arteriovenous malformations present within this time period.

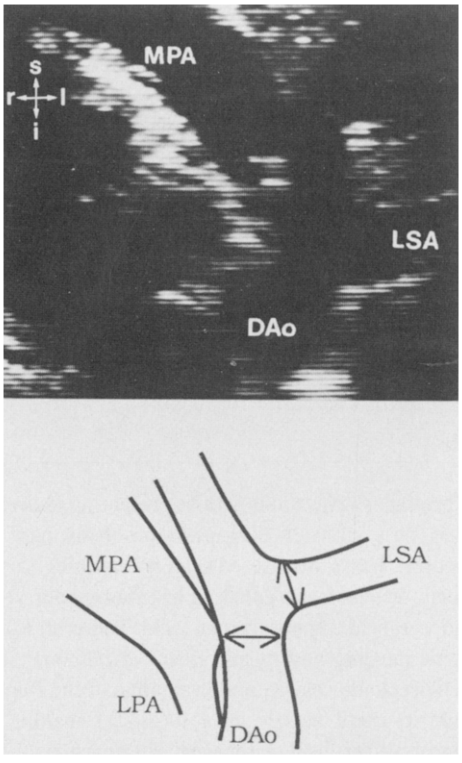

Figure 1. "Ductal view" of the aorta obtained from the high left parasternal area immediately below the left clavicle. In this view, the origin of the left subclavian artery (LSA), the isthmus and the descending aorta (DAo) immediately distal to the origin of the ductus arteriosus are visualized. Measurements of the left subclavian artery and descending aorta were taken at the designated sites on the schematic diagram. i = inferior; l = left; LPA = left pulmonary artery; MPA = main pulmonary artery; r = right; s = superior.

Echocardiographic evaluation. Echocardiograms in patients were examined for the following information: intracardiac anatomy, right versus left ventricular end-diastolic dimension measured from the parasternal short-axis view and dimensions of the ascending aorta immediately above the aortic valve sinuses, the transverse arch between the origins of the innominate and left common carotid arteries, the descending aorta immediately distal to the level of the left subclavian artery, the abdominal aorta at the level of the diaphragm and the dimensions of each of the brachiocephalic arteries immediately distal to their origins from the aortic arch. The right subclavian and common carotid arteries were measured immediately distal to their bifurcation. All echocardiograms were performed with the use of an ATL Mk 600 or Ultramark 8 (Advanced Technology Laboratories, Inc.) ultrasound system equipped with dual 7.5 MHz imaging and 5 MHz Doppler transducers.

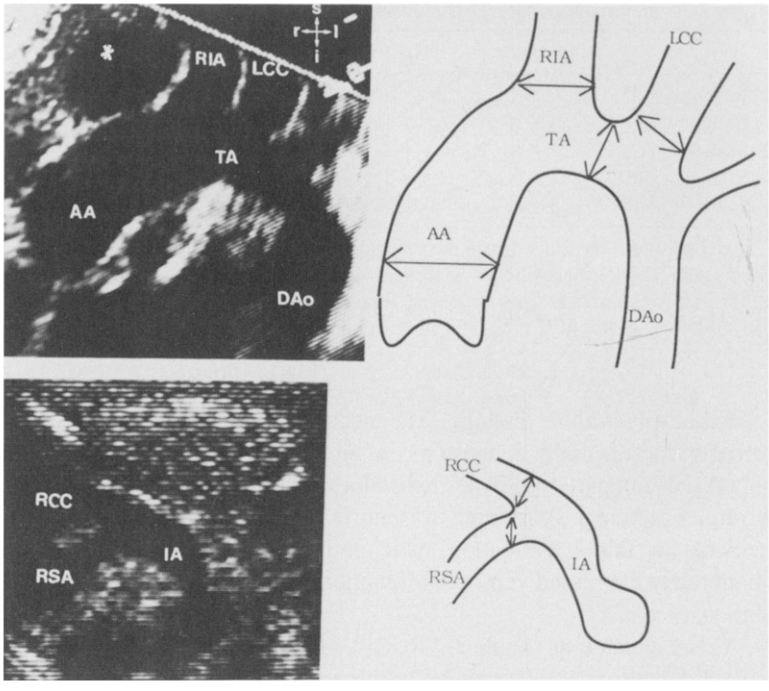

Figure 2. Cross-sectional view of the whole arch obtained from the standard suprasternal notch position (**upper**). The segments of the transverse aortic arch (TA) and the origins of the right innominate (RIA) and left common carotid (LCC) arteries were measured from this view at the sites shown in the schematic diagram. Note the enlarged innominate (IA) and left carotid arteries in this patient with a cerebral arteriovenous fistula. The dilated innominate vein is also seen superior to the transverse arch (*). The origins of the right common carotid (RCC) and right subclavian (RSA) arteries (image from a normal control infant) were measured from the view shown in the lower part of the figure obtained by angling the transducer superiorly toward the right shoulder from the frontal cross-sectional view of the aortic arch. AA = ascending aorta; DAo = descending aorta.

The high left parasternal view was used to evaluate the left subclavian artery and descending aorta (Fig. 1) and the standard suprasternal approach for the rest of the aortic arch and brachiocephalic vessels (Fig. 2). Selected good quality images showing the internal diameter of each vessel were acquired by a frame-grabbing system and measurements were performed on a digitizing pad.

Doppler spectral displays from each vessel were evaluated for the presence and direction of diastolic flow, defined as a separate spectral display after the return to baseline of systolic flow, or occurring after the T wave on the ECG with the signal to noise ratio optimized by wall noise filters set between 200 and 400 Hz. Doppler signals were obtained with an angle of incidence $\leq 20°$ in most cases. Prospectively acquired Doppler information in the control group was obtained at optimal angles. The left subclavian artery could not always be evaluated by Doppler signals at an angle $\leq 20°$ because of its position relative to the suprasternal view. However, because the ratio of diastolic to systolic flow and not absolute signal velocities was used in analysis, the angle of Doppler interrogation was only of relative importance. Systolic and diastolic Doppler spectral displays were digitized separately and the velocity-time integrals computed for comparison.

The size of the superior vena cava was subjectively assessed as normal or increased by comparing its diameter with that of the cross section of the aortic arch from the suprasternal frontal view. The quality of flow in the superior vena cava was also evaluated as normal phasic or turbulent nonphasic.

Statistical analysis. Measurements of the right and left ventricular end-diastolic dimensions in patients were compared with normal published values for infants. The abdominal aortic dimension in patients and control infants did not differ significantly and had a very small standard deviation (see Results). All other vessel dimensions were therefore expressed as ratios of the abdominal aortic dimension, and comparison of the mean values (± SD) for the ratio of each vessel to the abdominal aorta in patients and control infants was performed by the unpaired two-tailed *t* test. Absolute vessel dimensions were also compared with the unpaired two-tailed *t* test. A probability (p) value ≤ 0.05 was considered indicative of statistical significance.

Results

Clinical findings (Table 1). All patients presented in the first week of life and nine had signs of congestive heart failure. A cranial bruit was audible in five patients; however, in two of the five, it was heard only after definitive diagnosis by ultrasound or computed tomographic scanning. Ischemia on ECG, defined as ST segment depression of ≥ 1 mm or T wave inversion in the left precordial leads, was present in four of the seven patients with a cerebral arteriovenous fistula. Initial clinical diagnosis included various forms of congenital heart disease. One patient (Case 1) had diminished femoral pulses, and coarctation of the aorta was confirmed at angiocardiography, which also demonstrated a dilated innominate artery and rapid cerebral venous return.

Table 2. Comparison of Vessel Dimensions Between Patient and Control Groups

	Age (wk)	AA	TA	DA	IA	RSA	RCC	LCC	LSA	Abd A
Control group (n = 29)	10 ± 5	9 ± 1.5	7.3 ± 1.6	6.8 ± 1	4.5 ± 1	3.5 ± 1	4.2 ± 3	4.5 ± 1	3.6 ± 1	6.2 ± 1
Patient group (n = 10)	0.35 ± 0.3	11.5 ± 3	7 ± 1.5	5.5 ± 1.5	7.5 ± 2	5.2 ± 1.5	5.6 ± 2.5	5.3 ± 2.3	5.7 ± 2	6 ± 0.5
p Value	<0.0001	0.005	0.5	0.02	0.0001	0.003	0.4	0.03	0.007	0.83

All dimensions in mm. Although the patient group is significantly younger than the control group, the median age of the control group was 6 weeks (modal age 1 week). The differences in the vessel dimensions are thus even more significant because of the expected larger size of these vessels in the older control group. AA = ascending aorta; Abd A = abdominal aorta at level of diaphragm; DA = descending aorta; IA = innominate artery; LCC = left common carotid; LSA = left subclavian artery; RCC = right common carotid; RSA = right subclavian artery; TA = transverse arch.

Echocardiographic findings. The mean right and left ventricular end-diastolic dimensions in the 10 patients were increased compared with normal values for the same age group. Fractional shortening was normal (≥29%) in nine patients at initial evaluation. One patient (Case 7) had moderately depressed ventricular function when first seen at 7 days of age.

Vessel dimensions (Table 2). Results of measurements of vessel dimensions were rounded to the nearest 0.5 mm. The abdominal aortic dimensions in patients and control infants were 6.0 ± 0.5 and 6.2 ± 1 mm, respectively (p = NS). For this reason, vessel dimensions were expressed as ratios of the abdominal aorta. Comparison of this ratio between patients and control groups is shown in Figure 3. The general trend in both groups was for the vessel dimension to decrease from the ascending aorta toward the left subclavian artery, although all the vessels tended to be larger in the patient group. A notable exception to this observation was the descending aorta at the level of the left subclavian artery, which was significantly smaller in patients. The innominate artery in patients was equal to or larger than the transverse arch, with an innominate to transverse arch ratio of 1.2, versus 0.65 in the control group. In the two patients with a right subclavian arteriovenous fistula, the innominate and subclavian arteries were enlarged whereas the right carotid artery was not, in contrast, in those with a cerebral arteriovenous fistula, the innominate and right carotid arteries were enlarged and the right subclavian artery was normal. The site of fistulous communication between the subclavian artery and axillary vein was identified by pulsed Doppler ultrasound in one of these patients and later confirmed by digital subtraction angiography.

The superior vena cava was equal to or larger than the aortic arch cross-sectional diameter in six patients, including the two with a right subclavian arteriovenous fistula, whereas in the control group it was smaller than the aortic arch diameter. In one patient with a persistent left superior vena cava draining into the coronary sinus, both this vessel and the coronary sinus were markedly enlarged. Doppler

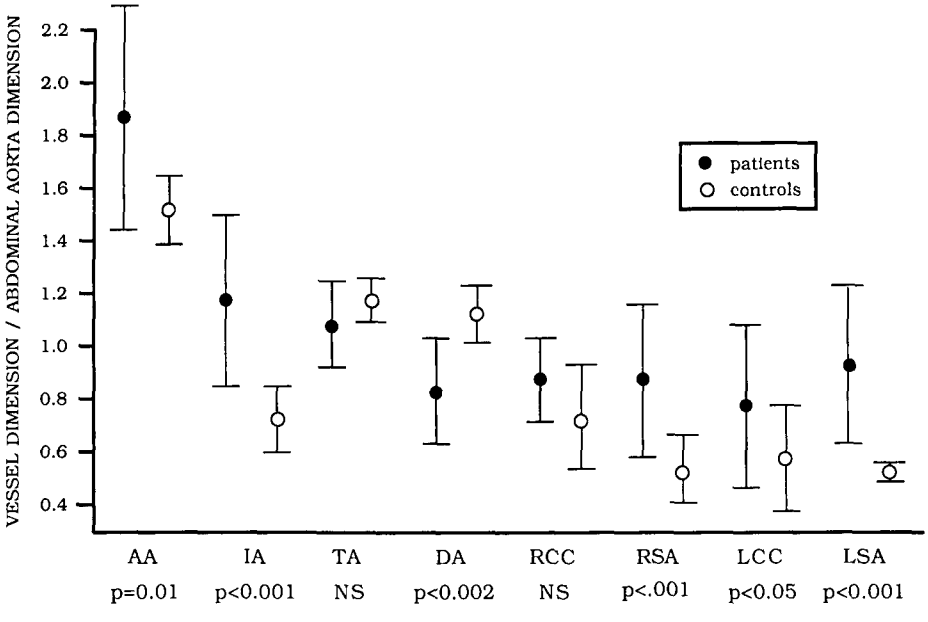

Figure 3. Aortic arch and brachiocephalic vessel dimensions expressed as ratios of the abdominal aorta measured at the level of the diaphragm. Dimensions in patients and control infants are denoted as means ± 1 SD. The p values refer to comparison of the ratios of the specified vessel to the abdominal aorta in patients and controls. AA = ascending aortia; DA = thoracic descending aorta at the level of the left subclavian artery; IA = innominate artery; LCC = left common carotid artery; LSA = left subclavian artery; RCC = right common carotid artery; RSA = right subclavian artery; TA = transverse arch.

Table 3. Diastolic Doppler Flow in Aortic Arch and Brachiocephalic Vessels in Patients and Control Groups

	AA	TA	DA	IA	RSA	RCC	LCC	LSA
Patient group (n = 10)								
No. with diastolic flow	2	6	6	4	3	1	3	1
Direction of diastolic flow	A	A5*	R	A	A	A	A	A
% Diastolic/systolic VTI	—		—	15%	45%	26%	27%	33%
Control group (n = 29)								
No. with diastolic flow	1	5	9	3	0	2	0	0
Direction of diastolic flow	A	A4*	A	A	—	—	—	—
% Diastolic/systolic flow	—	—	—	15%	—	14%	—	—

*Number of patients with antegrade diastolic flow; one each of the subjects in these groups had retrograde diastolic flow. A = Doppler diastolic flow in same direction as systolic flow (antegrade); R = Doppler diastolic flow in opposite direction to systolic flow (retrograde); VTI = Doppler spectral velocity-time integral; other abbreviations as in Table 2.

flow in the superior vena cava was characterized as turbulent in four patients.

Diastolic flow (Table 3). In the descending aorta proximal to the origin of the ductus arteriosus, six patients had retrograde and none had anterograde diastolic flow, whereas nine control infants had diastolic anterograde flow. Retrograde diastolic flow at this site was present only in patients. A typical diastolic flow profile in the aortic arch and innominate artery is shown in Figure 4. Aortic arch anterograde diastolic flow was present in the ascending aorta in 2 and in the transverse arch in 5 of the 10 patients, respectively, and in 1 and 4 of the 29 control infants, respectively. Retrograde diastolic flow was present in the transverse arch in one patient and one control infant.

Brachiocephalic vessel diastolic flow was always anterograde. When it was found in 5 of the 29 control infants, it was ≤15% of the systolic velocity-time integral. The diastolic/systolic velocity-time integral in the 10 patients ranged from 15 to 55% (mean 27). Specifically, diastolic antegrade flow >15% of systolic flow was present in every dilated vessel that was evaluated by Doppler ultrasound.

Correlation of vessels feeding the arteriovenous fistulas at cerebral angiography and vessels identified as being enlarged by echocardiography. As is usually the case, the

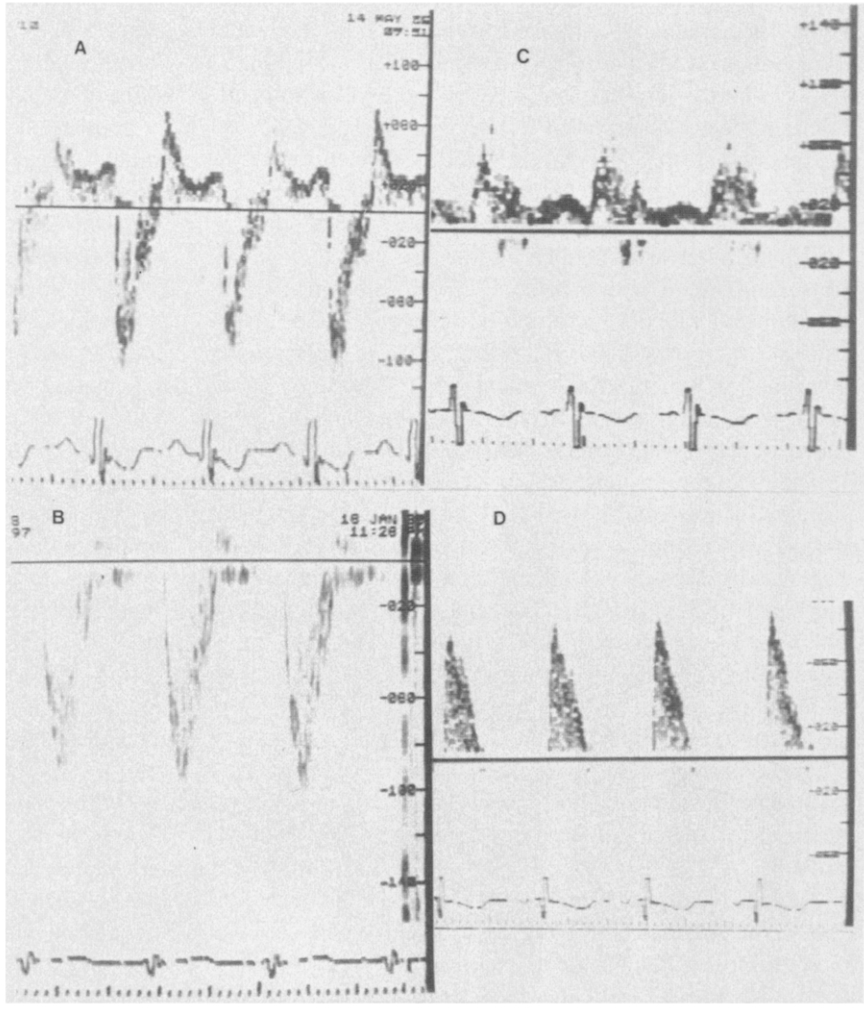

Figure 4. Left panels, Doppler spectral display in the descending aorta proximal to the ductus arteriosus in a patient (**A**) and control subject (**B**) obtained at the same gains and wall noise filter settings (200 Hz). Retrograde diastolic flow is demonstrated in the patient (**A**). **Right panels,** Doppler flow patterns from the right innominate artery in a patient (**C**) and control subject (**D**). The typical pattern of antegrade diastolic flow in the patient is shown. Flow toward the transducer is above the line.

majority of cerebral arteriovenous fistulas (six of seven) had multiple feeders arising from anterior or posterior cerebral and vertebral arteries with variations in which each vessel had major involvement. The right anterior cerebral artery was involved in six cases; the innominate artery was enlarged by echocardiography in these. Where there was dominant involvement of the vertebral artery, the left subclavian artery was markedly enlarged (n = 4) by echocardiography. There was thus excellent correlation between vessel enlargement by echocardiography and vessel involvement at cerebral arteriography.

Discussion

Our results show that imaging and measurement of brachiocephalic vessels, combined with Doppler flow evaluation, is a useful technique for the diagnosis of cerebral and thoracic arteriovenous fistulas in the newborn period. The finding of significant diastolic flow by Doppler ultrasound in vessels with an increased dimension can be used to localize the feeding vessel and direct further specific investigation.

Clinical diagnosis. Congestive heart failure was a common presentation in infants with cerebral arteriovenous fistulas in agreement with other reports (1,2). Systemic arteriovenous fistulas occurring at sites such as the liver and thorax, whereas less frequent, may also present with heart failure in the newborn period (1,3,4,5). In this series, one of two patients with a subclavian arteriovenous fistula presented in congestive heart failure. The correct diagnosis was not suspected in 5 of our 10 patients at initial clinical evaluation, whereas a cranial bruit was heard in only three of the seven patients with a cerebral arteriovenous fistula. In two of these (Patients 1 and 6), other diagnoses were given priority despite an audible cranial bruit because of the strong suspicion of structural congenital heart disease in both cases. This result serves to emphasize that although the finding of normal intracardiac anatomy in the setting of signs of heart failure in a newborn may lead one to suspect the presence of a cerebral arteriovenous fistula, the frequent absence of a cranial bruit (1) may result in erroneous diagnosis. Additionally, a cranial bruit of no clinical significance may be found in normal newborns, and a loud precordial murmur may be transmitted to the skull giving the false impression of a cranial bruit. Signs suggestive of coarctation of the aorta, which are not unusual in these infants (6), further add to the difficulties in diagnosis.

Angiocardiography, when carried out to confirm the diagnosis of a cerebral arteriovenous fistula, may contribute significantly to morbidity in these already critically ill infants.

Echocardiographic diagnosis. Attempts at noninvasive diagnosis of cerebral arteriovenous fistulas by echocardiography have thus far been confined to a few reports (3,7–9) describing dilated ventricular chambers and visualization of an "echo-free" intracranial space with the transducer placed on the anterior fontanelle. Use of air contrast echocardiography with detection of bubbles moving right to left across a patent foramen into the left ventricle and then returning within a few cardiac cycles by way of the superior vena cava was also briefly described in one study (8). However, this maneuver is not without risk. Specific evaluation of the aortic arch, including assessment of Doppler flow in these vessels, has not been reported.

In this study we carried out a quantitative analysis of the segments of the aortic arch and brachiocephalic vessels in 10 patients and compared these with measurements obtained in a group of 29 control infants using a methodology similar to that described by Morrow et al. (10). In all 10 patients, the ascending aorta and the innominate artery were significantly larger than those in the control group. Furthermore, when other brachiocephalic vessels were enlarged, these corresponded to the feeder vessels identified at cerebral angiography carried out before operative intervention in all patients. The accuracy with which echocardiographic imaging of the aortic arch reflects angiographic findings is well documented (11). The finding of increased ascending aortic and brachiocephalic vessel dimension is consistent with angiographic descriptions and is the result of increased blood flow through these vessels. Likewise, the smaller dimension of the descending aorta at the level of the subclavian artery in patients suggests diminished blood flow through the isthmus due to proximal run-off into the arteriovenous fistulas.

In this regard, this finding agrees with the theory of coarctation-like physiology in this entity (6) and mirrors the findings of Huhta et al. (11) in which quantitative echocardiographic assessment of the aortic arch segments in coarctation yielded results similar to ours with regard to relative hypoplasia of the proximal descending aorta. Although true coarctation of the aorta in association with cerebral arteriovenous fistulas is infrequent, it should always be sought.

Doppler diastolic flow with a mean spectral flow-time integral ≥15% of systolic flow was found in each dilated brachiocephalic vessel. Diastolic flow was found in the innominate artery in a small number of normal infants. In this setting, however, clinical diagnostic problems such as those in infants with arteriovenous fistulas are absent. This therefore should not detract from the value of diastolic flow as a specific finding indicating the presence of an arteriovenous fistula arising from a branch of the innominate artery.

Diastolic retrograde flow in the descending aorta proximal to the origin of the ductus arteriosus was found only in patients, even in the presence of a moderate patent ductus arteriosus in four. This flow pattern may also be found in association with severe aortic regurgitation or from where a large patent ductus has its origin from the innominate artery. Isolated severe congenital aortic regurgitation is rare, how-

ever, and a right-sided ductus is almost always found in association with abnormal intracardiac anatomy (12). The finding of retrograde diastolic Doppler flow in the proximal descending aorta in the setting of congestive heart failure with normal intracardiac anatomy should lead to careful evaluation of the size of the brachiocephalic vessels and the pattern of Doppler flow within them. Diastolic flow ≥15% of systolic flow in an enlarged brachiocephalic vessel should be followed by urgent definitive investigation because delay in diagnosis, resulting in progression of myocardial and cerebral ischemia and infarction, is implicated as one of the reasons for the high mortality in newborns and significant neurologic morbidity in survivors (13,14).

Conclusions. We have defined the dimensions of aortic arch segments and brachiocephalic vessels in a group of normal infants and the range of Doppler spectral display patterns in these vessels. In the setting of signs of heart failure in the newborn period, rapid noninvasive diagnosis of cerebral or thoracic arteriovenous fistulas can be reached by demonstrating increased dimensions of the ascending aorta and innominate artery in most patients, and various combinations of enlargement of the other brachiocephalic vessels, followed by definitive cranial ultrasound and cerebral angiography for confirmation.

Demonstration of diastolic Doppler flow may further aid in confirming the enlarged vessel as the origin of the fistulous communication. Evidence of enlargement of the superior vena cava and turbulent Doppler flow within it may be a helpful additional finding in some infants. With the advent of Doppler color flow, identification of abnormal systolic and diastolic flow within the brachiocephalic vessels should be easier, although our own preliminary experience suggests that problems with velocity aliasing and over sensitivity of some systems in encoding turbulence (which we have seen in normal infants) may be a source of potential problems. The need for careful pulsed Doppler interrogation of blood flow in these vessels, however, still remains important as the basis for a quantitative approach to evaluation of diastolic and systolic flow.

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